



Case Report

Nephrotic syndrome and pulmonary artery thrombosis

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ABSTRACT

Thromboembolic complications have emerged as a major hazard of the nephrotic syndrome. Thrombosis in both the arterial and venous sides of circulation has been reported. We report a case of a 22-year-old man who suffered pulmonary embolism involving the entire left lung and right ventrobasal segment.

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1. Introduction

Pulmonary embolism (PE) is a rare complication of nephrotic syndrome (NS). We present a case of PE involving the entire left lung and right ventrobasal segment. The patient was administered without thrombolytics and recovered completely.

2. Case report

A 22-year-old young man was transferred to our hospital because of cough and hemoptysis. He had been well until 3 months earlier, when facial and both lower extremities' edema gradually appeared. He was admitted to local hospital on 18 December 2003. Laboratory examinations revealed 4+ proteinuria, hypoalbuminemia (serum albumin = 15.3 g/L) and hyperlipidemia (triglyceride = 2.97 mmol/L, total cholesterol = 19.57 mmol/L). A diagnosis of NS was made. Two days later, the patient underwent a renal biopsy which revealed minimal change nephropathy. Prednisone, benazepril, simvastatin, dipyridamole and fluid restriction were administered to the patient. After 18 days of hospitalization, the patient's proteinuria resolved and he was discharged.

Forty days after discharge (Jan 16), the patient's proteinuria relapsed and he was readmitted to the same hospital. The same treatments were utilized. Two weeks later (Jan 30), the patient developed left-side chest pain exacerbating with breath. He had

fever, tachycardia and cough accompanying with blood-tinged sputa. He also felt chest distress and breathlessness after activity. A chest X-ray suspected pneumonia and left pleural effusion. Antibiotics were added. There was no improvement after 4 days' treatment and the level of serum albumin decreased to 11.1 g/L. Cellcept (0.5 g/d) was taken. A week later, there was still no obvious improvement and the patient had dipstick urine of 4+ protein. Therefore, the patient was referred to our hospital on 12 February 2004.

On admission, the temperature was 36.8 °C, the pulse 88 beats per minute, the blood pressure 100/60 mmHg, the respiratory rate 19 breaths per minute and no jugular venous distension. Serum albumin was 16 g/L and 24 h urinary protein excretion was 9.3 g. An electrocardiogram (ECG) showed no abnormal findings. Chest film revealed infiltration in the lower left lung and left pleural effusion. Initial diagnosis was made as pneumonia secondary to NS. The following chest CT revealed multiple patchy consolidations and an enlargement of pulmonary artery (Fig. 1). Considering NS is a risk factor for venous thromboembolism, further tests for suspected PE were done. Serum fibrinogen was 5.78 g/L (2–4 g/L). Thrombin time was 41.9 s (12–18 s). D-dimer test showed a negative result of 287 (<500 ng/mL). Antithrombin III (ATIII) level was 142 µg/L (<300 µg/L). Lung perfusion scan revealed total absence of perfusion to the left lung and slight nonuniform perfusion to the right ventrobasal segment (Fig. 2). Heparin infusion was continuously given at a dose of 200 U/Kg per day and the patient's condition gradually improved. Twenty days later, he was discharged. Half a year later, another lung perfusion scan confirmed no abnormalities. Followed up for four years, the patient recovered completely and now works in a ship manufacturer.

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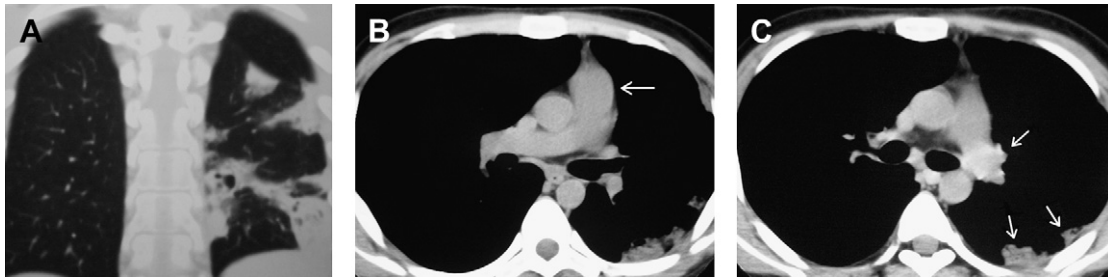


Fig. 1. A: Chest CT revealed multiple patchy consolidations in the left lung seem like triangle. B: The enlargement of the pulmonary arteries (arrow). C: Interrupted left pulmonary artery and triangular consolidations (arrow).

3. Discussion

The cumulative incidence of thromboembolic complications in patients with NS is nearly 50 percent. In adults with NS, arterial thrombosis is less common than venous thrombosis,¹ and such a large PE as described in this case is scarcely reported. Mechanisms for hypercoagulability in the NS include: (1) abnormalities in coagulation factors¹; (2) Glucocorticoids used stimulate bone marrow to produce more platelets; and (3) diuretics cause hypovolemia and pachyemia.

Glucocorticoids and immunosuppressive agents which are widely used in the treatment of NS often cause patients into a hyp immunity state. Symptoms such as cough, fever, leukocytosis and pleural effusion often mislead physicians to infectious diseases, for instance, pneumonia and tuberculosis.

On January 30, the patient had a moderate pretest probability for PE using the simplified Wells rule² due to the combination of immobilization, tachycardia and hemoptysis. The patient was misdiagnosed as pneumonia for nearly a month because PE was previously recognized as a rare disease in China which caused clinicians' lack of cognition and vigilance.

D-dimer is formed when cross-linked fibrin is lysed by plasmin, and elevated levels usually occur in patients with PE. But the result of D-dimer testing was negative in this case. After referring to the laboratory staffs, we were informed that the level of D-dimer was tested using an assay called immunoturbidimetry in our hospital. This method is less sensitive than enzyme-linked immunosorbent assay's (ELISA). We have experienced several cases of PE with negative D-dimer testing. So, it was suggested that the method used should be concerned when using the negative result of D-dimer to rule out PE. Lower sensitivity means higher probability of misdiagnosis which may means higher mortality. Meanwhile, if the clinical assessment is moderate or high, the D-dimer result needs to be combined with other examinations.³

Ventilation–perfusion lung scan has been the usual initial investigation in patients with suspected PE. When more than 3 segmental perfusion defects appeared, probability of PE is more than 90%.³ In this case, the patient's perfusion lung scan shows total absence of perfusion to the left lung and slight nonuniform perfusion to the right ventrobasal segment. It's a strong evidence to confirm the diagnosis. Pulmonary angiography is the gold standard, but it is expensive, invasive (mortality of about 0.5%) and

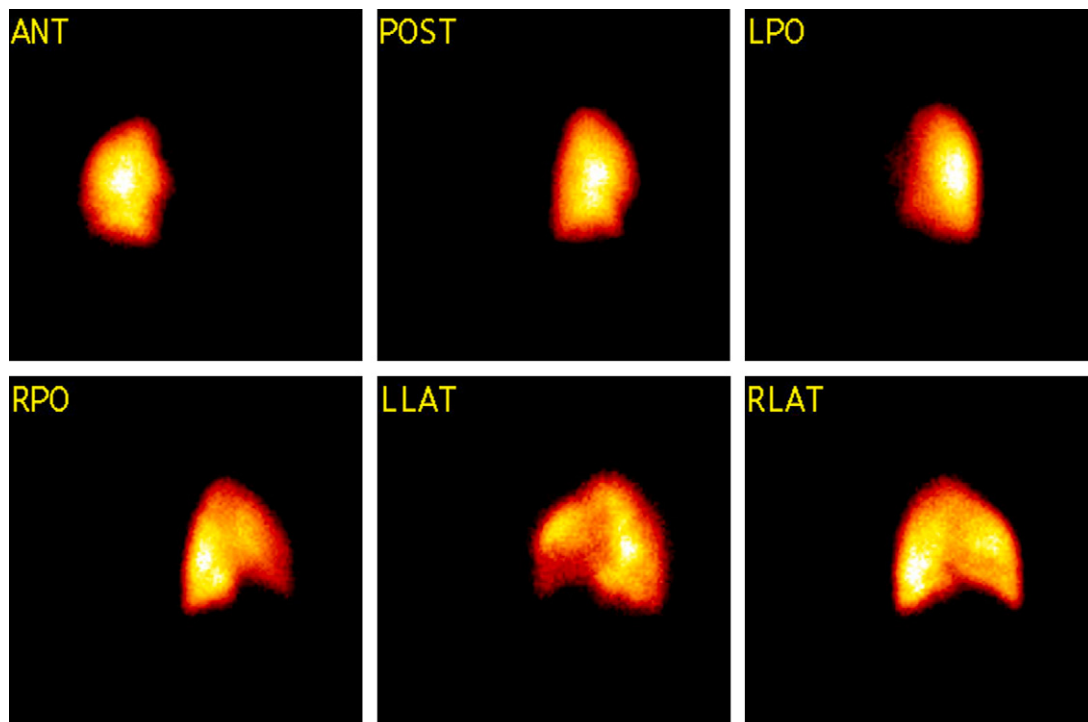


Fig. 2. Lung perfusion scan showing total absence of perfusion to the left lung and slight nonuniform perfusion to the right ventrobasal segment.

contraindicated in patients with renal impairment.³ Pulmonary angiography was not adopted because the result of perfusion was obvious and this poor patient had nephrosis.

During the whole course, the vital signs were generally stable and the ECG was normal. There were no obvious signs of hemodynamic change such as shock, which is rarely met in clinic. The reasons supposed may include: (1) it's a subacute PE, the right ventricle had time to adapt and became hypertrophy as pulmonary vascular resistance slowly increases and therefore could generate much higher pulmonary artery pressure; (2) left lung owns only 45% of pulmonary function. The lost can be compensated by the right lung; and (3) the patient was only 22-year-old and had no chronic pulmonary disease previously.

In general, thrombolytic therapy is indicated only for pulmonary embolism massive enough to threaten life or to leave the patient with inadequate pulmonary reserve. Most patients with lesions judged older than two weeks demonstrated little evidence of benefit from thrombolytic therapy.⁴ Considered 3% risk of a clinically significant hemorrhage,⁵ thrombolytics were not used. Anti-coagulants such as heparin and warfarin constitute the cornerstone of treatment for PE. This case proved that heparin alone might be effective for such a severe PE if there were no obvious signs of

hemodynamic change. The measurement of serum ATIII is also essential. If the level of ATIII reduced because of extra urinary loss caused by diuretics, heparin's antithrombotic effect will be diminished.⁶

Conflict of interest statement

The authors have no competing interests.

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